**Executive Summary**

**Purpose:** Ensure appropriate utilization and control of Diacomit® (stiripentol)

**Why Issue Selected:** In August 2018, the FDA approved Diacomit® (stiripentol) to be used in combination with clobazam for treatment of seizures associated with Dravet syndrome in patients 2 years of age and older. Dravet syndrome is a rare genetic condition that appears during the first year of life with frequent fever-related seizures. Later, other types of seizures typically arise, including myoclonic seizures. Additionally, status epilepticus, a potentially life-threatening state of continuous seizure activity requiring emergency medical care, may occur. Children with Dravet syndrome typically experience poor development of language and motor skills, hyperactivity and difficulty relating to others. Dravet syndrome has a higher mortality rate than other types of epilepsy, with most deaths occurring before 10 years of age. Dravet syndrome is estimated to appear in 1/15,700 births in the United States, or 0.0064% of the population. First line therapies for Dravet syndrome include clobazam, valproic acid, and cannabidiol. Diacomit is different from other seizure agents in that possible mechanisms of action include not only direct effects mediated through the gamma-aminobutyric acid (GABA)A receptor but also indirect effects involving inhibition of cytochrome P450 activity with resulting increases in blood levels of clobazam and its active metabolite.

**Program-Specific Information:**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Date Range FFS 01/01/2019 to 06/30/2019</th>
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<tbody>
<tr>
<td></td>
<td>Drug</td>
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<tr>
<td>Diacomit 250mg capsule</td>
<td>0</td>
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<tr>
<td>Diacomit 500mg capsule</td>
<td>0</td>
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<tr>
<td>Diacomit 250mg packet</td>
<td>0</td>
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<tr>
<td>Diacomit 500mg packet</td>
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**Type of Criteria:**
- ☒ Increased risk of ADE
- ☑ Appropriate Indications
- ☐ Preferred Drug List
- ☒ Clinical Edit

**Data Sources:**
- ☐ Only Administrative Databases
- ☒ Databases + Prescriber-Supplied
Setting & Population

- Drug class for review: Diacomit® (stiripentol)
- Age range: All appropriate MO HealthNet participants aged 2 years and older

Approval Criteria

- Participant aged 2 years or older AND
- Documented diagnosis of Dravet syndrome AND
- Concurrent use with clobazam AND
- Limit of 360 capsules or packets for the 250mg strengths and 180 capsules or packets for the 500mg strengths every 30 days AND
- For first claim only:
  - Documented trial of valproate (defined as 30 days in the past year) AND
  - Documented trial of Epidiolex (defined as 30 days in the past year) AND
  - Documented trial of clobazam (defined as 30 days in the past 60 days) AND
  - Prescribed by a neurologist or other appropriate specialist AND
  - Documentation of baseline neutrophil and platelet counts
- Renewal Criteria:
  - Initial approval of prior authorization is 3 months
  - Renewal of prior authorization may be up to 6 months following documentation of the following:
    - All approval criteria listed above
    - Lack of ADE/ADR to therapy
    - Complete blood count with differential required every 6 months

Denial Criteria

- Therapy with be denied if no approval criteria are met

Required Documentation

- Laboratory Results: X
- Progress Notes: X
- MedWatch Form: 
- Other: 

Disposition of Edit

Denial: Exception “682” (Clinical Edit)

References

- Diacomit [package insert]. France: Biocodex; August 2018